

Subtle clinical signs of a spinal cord ependymoma at the cervicothoracic level in an adult: a case report

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A 33-year-old male presented to a chiropractic clinic complaining of chronic, recurrent low back pain. Subtle signs of muscle atrophy were noted in the left hand during the history taking. This muscle atrophy was reported as having a gradual onset spanning the past six months without any precipitating event. Cervical, thoracic and lumbar spinal radiographs were deemed unremarkable. Due to the progressive nature of the neurological deficit, the patient was referred for a neurological consultation. A magnetic resonance imaging (MRI) study was performed and revealed an expansive intramedullary lesion between C6 and T1 suggesting a differential diagnosis of spinal cord ependymoma or astrocytoma. The patient underwent surgical excision of the tumour. Pathological report confirmed a diagnosis of ependymoma. In the presence of subtle clinical signs, clinicians should keep a high index of suspicion for spinal cord tumours. (JCCA 2006; 50(4):244–248)

KEY WORDS: ependymoma, tumour, spinal cord, cervicothoracic.

Introduction

In a population-based survey of 467 patients with primary intraspinal neoplasms, intramedullary ependymomas accounted for 34.5% of all ependymomas of the central nervous system.¹ According to the same study, the age-adjusted incidence rate for the primary intraspinal neo-

Un homme de 33 ans s'est pr sent    une clinique de chiropratique se plaignant de douleurs chroniques et r currentes au bas du dos. Au cours de l'interrogatoire, on a observ  de subtils signes d'atrophie musculaire   sa main gauche. Cette atrophie se serait graduellement install e au cours des six derniers mois sans qu'il y ait eu d' v nement d clencheur. Des radiographies de la nuque, du thorax et de la moelle  pini re lombaire n'ont rien r v l  d'anormal. En raison de la nature progressive du d ficit neurologique, on a r f r  le patient pour une consultation en neurologie. Une  tude d'imagerie par r sonance magn tique (IRM) a  t  effectu e et a r v l  une l sion centrom dullaire en progression entre C6 et T1, ce qui nous a amen    opter pour un diagnostic diff rent d' pendymome de la moelle  pini re ou astrocytome. On a proc d    une excision de la tumeur. Le rapport pathologique a confirm  le diagnostic d' pendymome. En pr sence de signes cliniques subtils, les cliniciens devraient garder frais   la m moire la possibilit  qu'il s'agisse d'une tumeur de la moelle  pini re. (JACC 2006; 50(4):244–248)

MOTS CL S :  pendymome, tumeur, moelle  pini re, cervico-thoracique.

plasm is 0.5 in females and 0.3 in males per 100,000 population per year. Although reported in all age groups, intraspinal ependymomas are more frequently seen in the adult population.² Some authors believe it is more common in the fourth and fifth decade while others propose a wider distribution spanning between the second and sixth

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decade of life.^{2,3,4,5,6} While a male predominance has been suggested, other authors have found equal proportions between genders.^{1,4,7}

We present a case of a 33-year-old male with a cervico-thoracic ependymoma. The objective of this case report is to demonstrate uncommon characteristics of a rare condition in a patient that may present to chiropractors or other health care professionals.

Case report

A right handed 33-year-old male presented to a chiropractic clinic complaining of chronic recurrent low back pain. Subtle signs of left hand muscle atrophy were noted during the history taking. Upon further questioning, the patient reported this had gradually appeared over the past six months and was accompanied by a mild loss of grip strength. He had no previous history of cervical, thoracic or brachial pain, paresthesia, cramping or fasciculation. He reported no complaints in the other limbs and no bowel or bladder dysfunction. His past medical history and family history were unremarkable. He considered himself in general good health.

On physical examination, cervical and lumbar spine active and passive ranges of motion were full and pain free. Neurological testing of the upper and lower extremities revealed left hand motor weakness, rated 3/5. Weakness was noted with abduction, adduction and flexion of the fingers corresponding to the C8–T1 myotome. Sensory response was normal and deep tendon reflexes (DTR) (biceps, brachioradialis and triceps) were graded +2 bilaterally. Heel and toe walking and Romberg's test were unremarkable. Although no clonus was noted, plantar response was deemed equivocal on the left side.

The patient was referred to his medical doctor who ordered antero-posterior (AP) and lateral radiographs of the cervical, thoracic and lumbar spine as well as oblique views of the cervical spine. All films were read as normal.

Due to the progressive nature of the neurological deficit, the patient was referred for a neurological consultation which took place two months after his initial presentation to the chiropractor.

Intervention and outcome

The neurologist's physical examination revealed weakness in the left upper limb that had progressed over the past two months. The atrophy and the weakness of the

abductor pollicis brevis and interosseous muscles had also increased. The DTR of the triceps and flexor digitorum muscles were absent bilaterally.

An electromyography (EMG) study was performed followed by magnetic resonance imaging (MRI). The results of the EMG study suggested the presence of a chronic left C8 radiculopathy with active denervation. The MRI study revealed an expansive intramedullary lesion with peripheral enhancement and extending from C6 to T1 suggesting a differential diagnosis of spinal cord ependymoma or astrocytoma. No osseous abnormalities were detected (Figures 1 and 2).

The patient underwent near complete surgical excision of the tumour. The pathological report confirmed a diagnosis of an ependymoma.

The patient was recommended to attend an intensive physical rehabilitation program. At four months post-surgical follow-up, neurological examination revealed mild gait unsteadiness likely due to a loss of posterior column proprioception. Motor power testing and muscle atrophy of the left hand were unchanged while right arm evaluation was normal. He started jogging again at six month post-surgery and attended his first marathon a year later. He was seen periodically by the chiropractor for upper back stiffness and was treated by gentle spinal manipulation of the thoracic spine and soft tissue therapy of the cervical spine. Left hand motor strength had partially returned while the muscle atrophy persisted. He was left with a mild subjective sensory deficit of the left leg.

Discussion

"Tumors of the spine may arise from the neural tissue, the meninges, the bone or surrounding soft tissues, or embryonal rests or as a result of derangement of embryogenesis, or they may spread from other tumors to the spine".² Meningioma is the most common primary intraspinal neoplasm, followed by glioma and neurilemmoma.¹ Meningioma and neurilemmoma are generally described as an extramedullary but intradural mass.² "Gliomas are the predominant type of intramedullary tumor. Subclassification includes ependymoma, astrocytoma, oligodendroglioma, glioblastoma and medulloblastoma".² Ependymoma is reported as the most common subtype, accounting for 2–8% of all primary central nervous system tumours.^{2,8} Ependymomas are also the third most common brain tumour in kids.⁹ Typically found in



Figure 1 Expansive intramedullary lesion with peripheral enhancement and extending from C6 to T1. Sagittal MR image (TR:3000, TE:110).

the infratentorial region in children, ependymomas tend to be located in the supratentorial region in young adults.^{3,4,7,9}

The precise tumour distribution of intraspinal ependymomas remains unclear, although predominance for the cervical region has been suggested. In small sample size studies,^{4,5,7} spinal cord ependymomas were located in terminal filum and/or cauda equina regions (31–45%) followed by cervical spine region (13.2–40%), lumbar region (39.6%), thoracic region (11.5–29%), cervicothoracic region (11.3–19.2%) and thoracolumbar region (15.4–20.8%).

Ependymomas originate from the ependyma cells, which are lining the spinal cord's central canal.¹⁰ "These cells vary from cuboidal to flat and modulate the transfer of fluid between the cerebrospinal fluid (CSF) and the cells

of the nervous system".¹¹ The pathognomonic feature of the ependymoma's cells is their tendency to encircle blood vessels (perivascular pseudorosettes). Ependymoma's are typically slow growing and can obstruct the CSF flow as well as compress centrally the spinal cord.¹⁰

Primary symptoms of spinal cord ependymomas include neck and back pain, often corresponding with the level of the spinal cord lesion.¹² While shoulder and interscapular pain seem more common in cervical and cervicothoracic tumour cases, patients with ependymomas of the cauda equina region may report lower back, leg and sacral pain mimicking lumbar disc disease.^{12,13} Pain is the initial presenting complaint for the majority of patients (65%) and may be present for a long period prior to the onset of neurological signs and symptoms, that is, for an average of 16 months.¹² Associated clinical

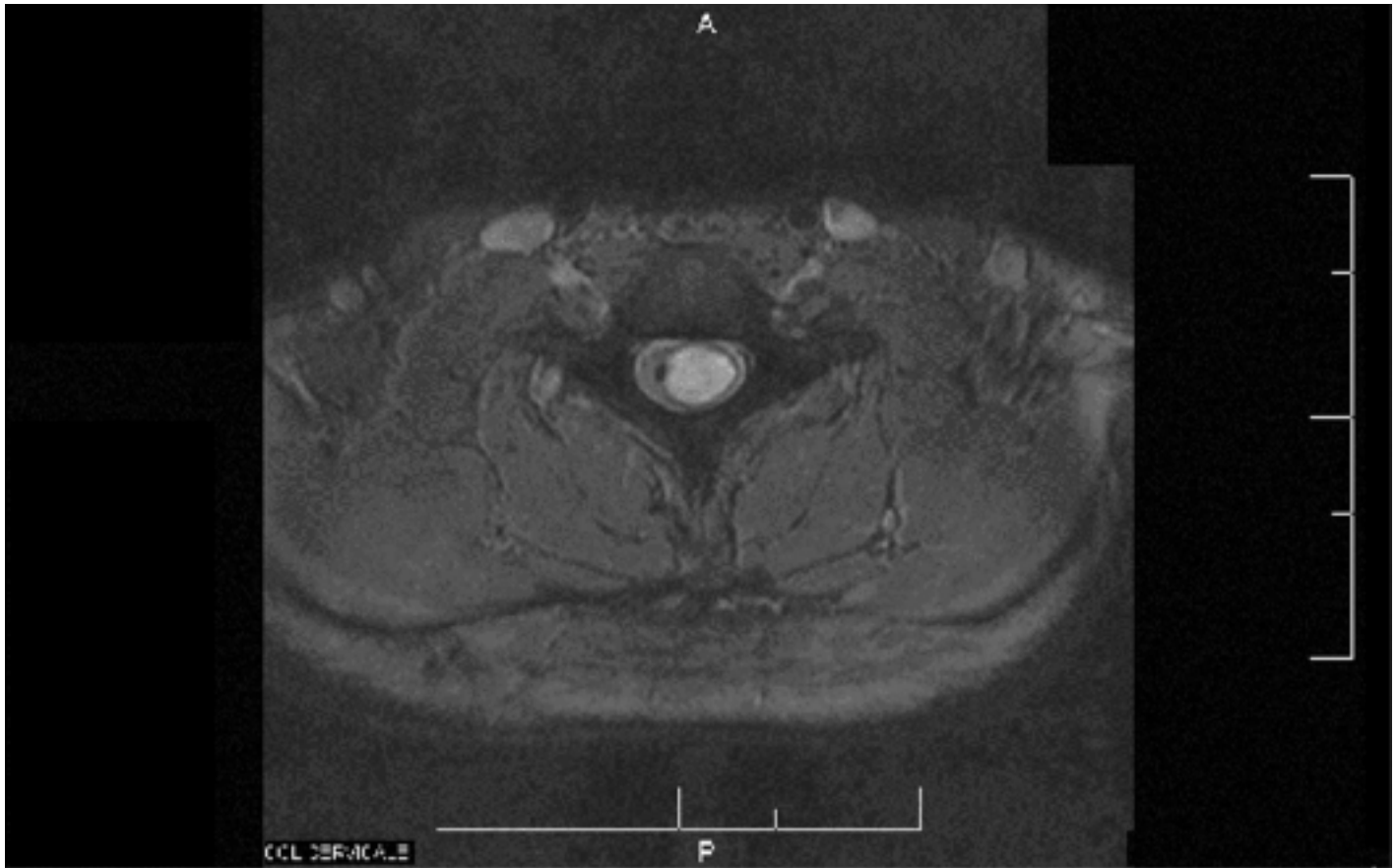


Figure 2 Compression of the spinal cord. Transaxial 3D-volume acquisition, gradient echo MR image (TR:50, TE:24).

manifestations include radicular pain, unsteady gait, numbness, paresthesia and bladder/bowel dysfunction. Thoracic and lumbar intraspinal neoplasms may present with distal numbness of the lower extremities and progress proximally. Sensory findings, in particular dysesthesias, are present in 70% of the patients.⁶

Examination of the cervicobrachial region may reveal weakness and/or atrophy of one or both arms, proximal muscle atrophy and incomplete Brown-Séquard syndrome. McCormick, Torres, Post and Stein,¹² reported unilateral or bilateral hand atrophy as a common sign of cervical intraspinal tumours. In their study, two other patients presenting with thoracic ependymomas initially experienced legs numbness and only after a delay of fourteen months and two years respectively did they show some mild leg weakness and stiffness. One developed bowel and bladder dysfunction. In another report,¹³

53% of subjects suffering from cauda equina ependymomas presented either with pain alone in the lower back, legs or sacral region, or with pain and paresthesia. Another 26% of patients had both pain and leg weakness or diminished reflexes. The remaining 20% had a combination of pain, weakness and sphincter dysfunction. Due to the small sample size in most of the available studies, it is not possible to relate the tumour stages to the clinical presentation. On average, there appears to be a delay of one to four years between the onset of symptoms and the time of diagnosis, mostly because of the slow tumour growth of ependymomas.^{5,6,12,13} L'hermitte's sign, a "lightening-like" sharp pain radiating from the base of the neck down to the back or distally in the extremities, lasting only seconds may occasionally be present.¹⁴

Plain film radiographs of the spine may show abnormal interpedicular space widening, thinning of the lami-

na, and erosion of the posterior elements (scalloping of the vertebral body) or of the medial aspect of the pedicle. Even so, many radiographic studies are unremarkable. Diagnostic imaging can include myelogram, CT-myelogram or magnetic resonance imaging (MRI) which typically demonstrate intermediate to hyperintense signal on T2 weighted images and isointense to slightly hypointense signal on T1 weighted images.^{3,4,12,13}

There is also a paucity of information with regard to laboratory findings. One case series³ reported elevation of cerebrospinal fluid protein values in 10 out of 19 patients with spinal cord ependymomas.

Appropriate treatment protocol remains somewhat controversial. Most authors suggest total tumour resection when feasible. However, partial resection is recommended if additional function may be lost.⁶ Subsequent treatments may include radiation and/or chemotherapy but there is currently no consensus about the success rate of such treatment modalities.¹⁵ Patients with better survival rates are those who had complete resection of a spinal ependymoma when compared to those with brain ependymoma in the absence of metastases.^{3,8,16} According to several authors, recurrence is more common in cases of sub-total tumour resection, but others have not reported any significant difference.^{3,5,8} The proposed incidence of metastasis also varies. In a review on ependymoma epidemiological and clinical features,⁸ the composite rate (among published series) of metastasis for spinal ependymomas was 3.7%. However, the type of therapy patients underwent is unknown. In accordance with other studies, Asazuma, Toyama, Suzuki, Fujimura and Hirabayshi⁵ reported "the overall 5-year survival rate for spinal ependymomas to be between 57% and 90%". Large scale studies are necessary in order to gain additional insight into ependymoma tumours.

Conclusion

Due to the rarity of this tumour, it is difficult to draw any conclusions regarding clinical presentation, tumour description, treatment, survival rate, incidence of recurrence and metastasis with either total or sub-total resection, with or without additional therapy. In the presence of subtle clinical signs such as those presented in this case report, clinicians should keep a high index of suspicion for spinal cord tumours. Specifically, in the presence of persistent or progressive neurological signs and symptoms, sphincter

dysfunction, or failed conservative therapy, clinicians should not hesitate to refer the patient for further evaluation as part of a complete management program.

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